

Intramastoid solitary neurofibroma mimicking middle ear carcinoma

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Abstract

We report an extremely rare case of intramastoid neurofibroma. A mass with destruction of the mastoid bone of a 51-year-old woman was examined with computed tomography. Subsequent magnetic resonance imaging demonstrated an ill-defined soft tissue mass with the opacification of mastoid air cells that had a mass effect in the same area. The patient underwent left subtotal temporal bone resection, and histological and immunohistochemical findings confirmed the lesion to be a neurofibroma. Given that similar imaging features of neurofibroma have been reported previously elsewhere in the head/neck and extremities, we suggest that it may be possible to include this tumor in the preoperative differential diagnosis.

Keywords

Magnetic resonance imaging (MRI), neurofibroma, mastoid bone, destruction, computed tomography (CT)

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Introduction

Solitary neurofibromas are common benign peripheral nerve sheath tumors and occur in patients without neurofibromatosis type 1. Although the head and neck region is a common location for benign peripheral nerve sheath tumors, intramastoid solitary neurofibromas are exceedingly rare. To our knowledge, 21 cases of intra-temporal solitary neurofibromas have been reported in the literature (1–4). Only one case of intramastoid solitary neurofibroma has been reported in English and Chinese literature (5). We present a case of intramastoid solitary neurofibroma, and we review the radiological, gross histological, and immunohistochemical features.

Case report

A 51-year-old woman with a 3-year history of left-sided otalgia underwent computed tomography (CT). A 2.67×1.86 cm ill-circumscribed tumor-like mass with temporal bone destruction was found (Fig. 1). The mass spread into the external auditory canal and middle ear. Internally, the soft tissue mass demonstrated heterogeneous density, in the range of 31–49 Hounsfield units, incompatible with an irregular, “moth-eaten” bone destruction pattern on high-resolution computed tomography images (Fig. 2).

Magnetic resonance imaging (1.5T Signa, GE Healthcare, Milwaukee, WI, USA) with contrast medium enhancement, which was performed for further characterization, showed a 2.8×2.4 cm T2 slightly hyperintense mass in comparison to that of the adjacent sternocleidomastoid muscle (Fig. 3). On T1-weighted (T1W) images, the mass showed slightly hypointense signal intensity with irregular shape (Fig. 4). On fat-suppressed water-sensitive sequences, the tumor showed homogeneous high signal intensity. The tumor showed high signal intensity on DWI-EPI sequence (b value = 1000 s/mm^2) and low signal intensity on ADC map, confirmed to represent restricted diffusion of the lesion (ADC value: $1.02 \times 10^{-3} \text{ mm}^2/\text{s}$). The mass showed strong and homogenous contrast

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Fig. 1. Axial CT shows a tumor-like mass (arrow) spreading into the external auditory canal.

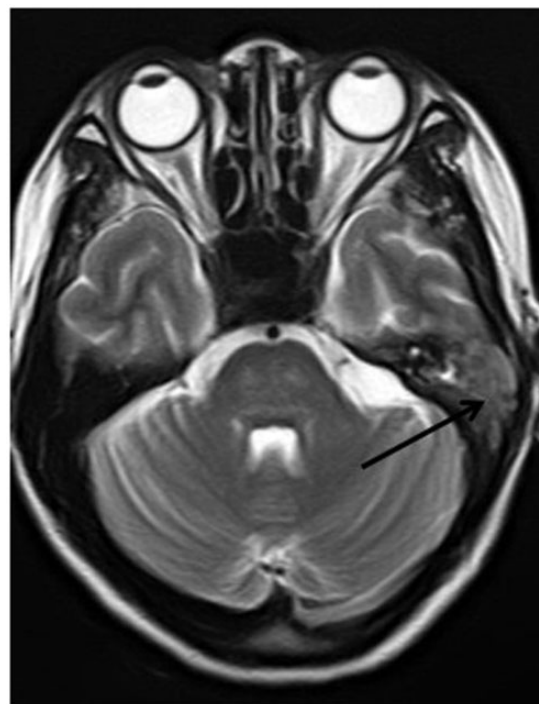


Fig. 3. Axial FSE T2W MR image shows a homogeneous T2 hyperintense mass (arrow) occupying the left mastoid air cells and extending into the external auditory canal and middle ear.

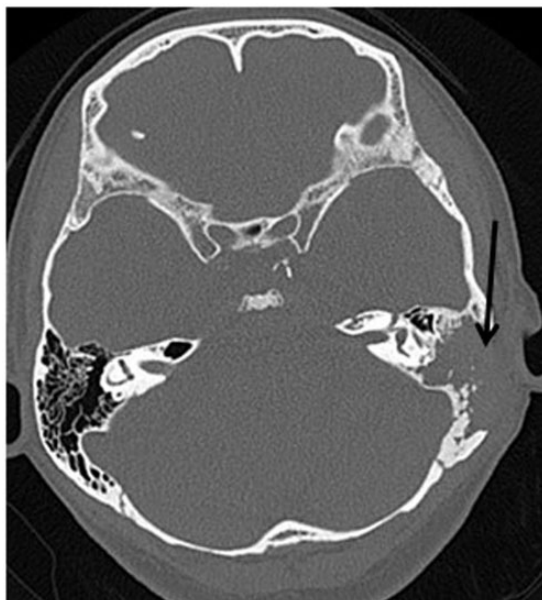


Fig. 2. High-resolution CT shows "moth-eaten" bone destruction pattern (arrow) within the mastoid.

medium enhancement on T1W fat-suppressed MRI (Fig. 5). The lesion showed no central or peripheral necrosis and no cystic formation. There was no associated invasion of the adjacent sternocleidomastoid muscle, intracranial extension, lymphadenopathy, or vascular encasement. After CT and MRI, the mass

remained non-specific and a malignant tumor could not be excluded. Diagnostic considerations included malignant tumors known to originate within the external auditory canal and middle ear, as squamous cell carcinoma or adenoid cystic carcinoma, and an unusual appearance of malignant peripheral nerve sheath tumors, although there was no cystic formation and ring-enhanced pattern to suggest the latter diagnoses strongly. No other head and neck mass was identified. Physical examination did not show any *café au lait* spots or other cutaneous or mucosal stigmata that would strongly have suggested neurofibromatosis type 1.

The patient underwent left subtotal temporal bone resection. The tumor was located within the mastoid bone, extended into the external auditory canal and middle ear. Microscopic examination of the tumor revealed interlacing bundles of spindle cells with wavy, dark-staining nuclei and destructive bone (Fig. 6).

Cells that had wavy nuclei were associated with abundant amounts of collagenous background. Mast cells, lymphocytes, and a small number of perineurial cells were dispersed throughout the stroma. The cutaneous appendages were involved. No Antoni A cells, typical of a Schwannoma, were seen. No atypia was noted and mitotic figures were rare.

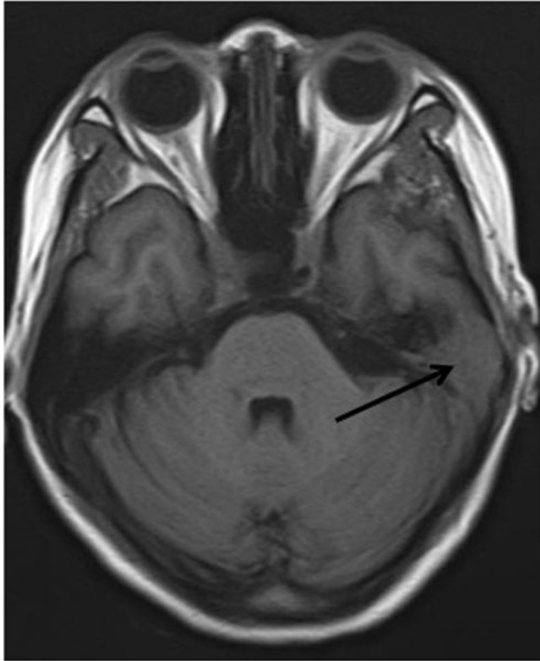


Fig. 4. Axial FSE T1W MR image shows a homogeneous T1 hypointense mass (arrow) occupying the left mastoid air cells.

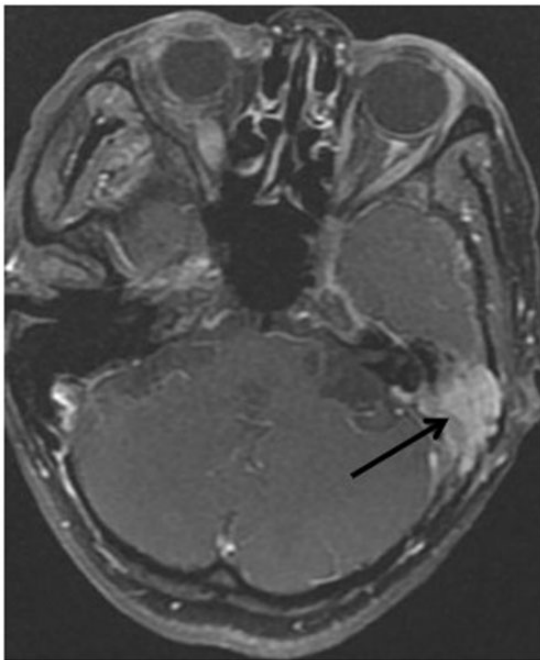


Fig. 5. The mass is enhanced strongly and wholly on contrast-enhanced T1W MR image.

The neoplastic cells were positive for S-100, CK, EMA, SMA, P63, CD3 (cluster of differentiation 3), CD20 on B cells, and negative for desmin and CD68. On the basis of these immunohistochemical and

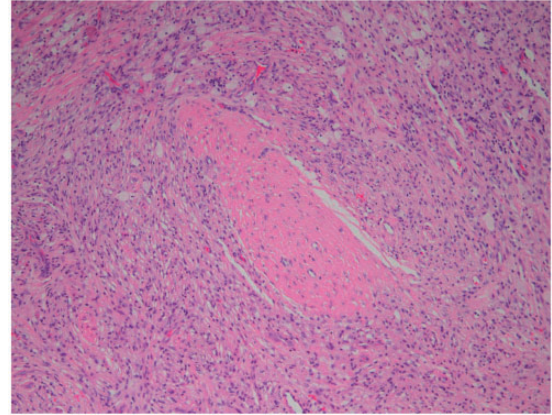


Fig. 6. Section from the tumor demonstrating interlacing bundles of spindle cells with wavy, dark-staining nuclei in a collagenous stroma (hematoxylin-eosin, original magnification $\times 100$).

histological features, the solitary tumor was diagnosed as a neurofibroma.

Discussion

Solitary neurofibromas are comparatively rare and usually found within cutaneous or subcutaneous tissues or along nerves (6) and represent 5% of all benign soft tissue neoplasms. Although the head and neck region is a common location for benign peripheral nerve sheath tumors, the occurrence of intramastoid neurofibromas is particularly uncommon. They are more likely to occur within the salivary gland rather than within mastoid itself (7). When such a diagnosis is suspected, a frozen section analysis may be considered at the time of surgery to establish both the benign nature of the tumor, thereby assisting the avoidance of unnecessary subtotal temporal bone resection. Although the rate of malignant degeneration of neurofibromas in patients with neurofibromatosis type 1 has been estimated at 2–29%, the rate of malignant transformation of solitary neurofibromas is believed to be extraordinarily low (8). In addition, local recurrence rate of solitary neurofibromas is low if the lesion is completely excised (9).

This case involved a solitary intramastoid neurofibroma extended into the external auditory canal and middle ear. The occurrence of a solitary neurofibroma within the temporal bone is rare (3), with only 21 such tumors previously reported in the literature to our knowledge. The differential diagnosis includes acute otomastoiditis, a facial nerve Schwannoma, or malignant mastoid bone invasion such as squamous cell carcinoma and adenoid cystic carcinoma of the head and neck. All of these tumors can be located within or

spread into the mastoid bone and produce the destruction of the temporal bone. Despite their similar anatomical location, the clinical presentations, and the CT and MRI appearance of the tumors differ. Acute otomastoiditis is common in children who present with fevers, otalgia, otorrhea, and retro-auricular swelling. At CT, there is opacification of the tympanic cavity and/or mastoid, with possible fluid levels and temporal bone destruction (10). Schwannomas of the facial nerve are also rare. CT and MRI findings related to Schwannoma of the head and neck are similar to those of neurofibroma and cannot be distinguished in many cases. However, some target imaging features can help differentiate these two lesions. A heterogeneous appearance with degeneration and cavitation on MRI image is much more common in Schwannoma than in neurofibroma (11). Multiple small ring-like hyperintense structures on T2-weighted (T2W) images and water-sensitive sequences, referred to as a “fascicular appearance” are also more commonly found in Schwannoma than in neurofibroma. The detection of a capsule on MRI could be useful in differential diagnosis because Schwannoma is an encapsulated mass unlike neurofibroma.

Squamous cell carcinoma can produce focal or diffuse contrast-enhancing soft tissue with bone destruction of the temporal bone; however, skin ulceration can commonly be seen on physical examination, often with local nodal metastases. ADC value can play an important complementary help in differential diagnosis. Some authors propose ADC values of the benign tumors were higher than that of the malignant tumors (12).

Our report is the first to show the CT and MRI appearance of an intramastoid neurofibroma. Review of the CT images from five earlier reports (1–5) of this lesion shows a similar imaging appearance, a soft tissue shadow was observed.

In conclusion, non-specific CT and MRI imaging features, resection and histopathological analysis remains necessary to establish the exact diagnosis. Percutaneous biopsy is necessary.

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